Complete Summary

GUIDELINE TITLE

Dilatation of the upper urinary tract (ureteropelvic junction and ureterovesical junction obstruction). In: Guidelines on paediatric urology.

BIBLIOGRAPHIC SOURCE(S)

Dilatation of the upper urinary tract (ureteropelvic junction and ureterovesical junction obstruction). In: Tekgul S, Riedmiller H, Gerharz E, Hoebeke P, Kocvara R, Nijman R, Radmayr C, Stein R. Guidelines on paediatric urology. Arnhem, The Netherlands: European Association of Urology, European Society for Paediatric Urology; 2008 Mar. p. 44-7. [15 references]

GUIDELINE STATUS

This is the current release of the guideline.

COMPLETE SUMMARY CONTENT

SCOPE

DISEASE/CONDITION(S)

- Ureteropelvic junction obstruction
- Ureterovesical junction obstruction (primary obstructive megaureter)
- Secondary megaureter
- Neonatal hydronephrosis

GUIDELINE CATEGORY

Counseling
Diagnosis
Evaluation

Management
Screening
Treatment

CLINICAL SPECIALTY

Obstetrics and Gynecology
Pediatrics
Surgery
Urology

INTENDED USERS

Physicians

GUIDELINE OBJECTIVE(S)

- To outline a practical and preliminary approach to paediatric urological problems
- To increase the quality of care for children with urological problems

TARGET POPULATION

Newborn infants and children with dilatation of the upper urinary tract

INTERVENTIONS AND PRACTICES CONSIDERED

Diagnosis

1. Antenatal and postnatal ultrasound
2. Voiding cystourethrogram
3. Diuretic renography

Treatment/Management

1. Prenatal counseling
2. Intrauterine intervention (rarely recommended)
3. Management of ureteropelvic junction obstruction
    - Symptomatic: pyeloplasty
    - Asymptomatic: conservative management
4. Management of secondary megaureter
    - Conservative management
    - Surgical reconstruction/antireflux repair

MAJOR OUTCOMES CONSIDERED

- Prenatal detection rate of reflux
- Incidence of neonatal hydronephrosis
- Rate of spontaneous remission

METHODOLOGY

METHODS USED TO COLLECT/SELECT EVIDENCE

Searches of Electronic Databases

DESCRIPTION OF METHODS USED TO COLLECT/SELECT THE EVIDENCE

The guidelines were based on current literature following a systematic review using MEDLINE.

NUMBER OF SOURCE DOCUMENTS

Not stated

METHODS USED TO ASSESS THE QUALITY AND STRENGTH OF THE EVIDENCE

Weighting According to a Rating Scheme (Scheme Given)

RATING SCHEME FOR THE STRENGTH OF THE EVIDENCE

Levels of Evidence

1a Evidence obtained from meta-analysis of randomized trials

1b Evidence obtained from at least one randomized trial

2a Evidence obtained from at least one well-designed controlled study without randomization

2b Evidence obtained from at least one other type of well-designed quasi-experimental study

3 Evidence obtained from well-designed non-experimental studies, such as comparative studies, correlation studies and case reports

4 Evidence obtained from expert committee reports or opinions or clinical experience of respected authorities

METHODS USED TO ANALYZE THE EVIDENCE

Systematic Review

DESCRIPTION OF THE METHODS USED TO ANALYZE THE EVIDENCE

Application of a structured analysis of the literature was not possible due to a lack of well-designed studies. Whenever possible, statements have been classified in terms of level of evidence and grade of recommendation. Due to the limited

availability of large randomized controlled trials – influenced also by the fact that a considerable number of treatment options relate to surgical interventions on a large spectrum of different congenital problems – this document is therefore largely a consensus document.

METHODS USED TO FORMULATE THE RECOMMENDATIONS

Expert Consensus

DESCRIPTION OF METHODS USED TO FORMULATE THE RECOMMENDATIONS

- The first step in the European Association of Urology (EAU) guidelines procedure is to define the main topic.
- The second step is to establish a working group. The working groups comprise about 4-8 members, from several countries. Most of the working group members are academic urologists with a special interest in the topic. In general, general practitioners or patient representatives are not part of the working groups. A chairman leads each group. A collaborative working group consisting of members representing the European Society for Paediatric Urology (ESPU) and the EAU has gathered in an effort to produce the current update of the paediatric urology guidelines.
- The third step is to collect and evaluate the underlying evidence from the published literature.
- The fourth step is to structure and present the information. The strength of the recommendation is clearly marked in three grades (A-C), depending on the evidence source upon which the recommendation is based. Every possible effort is made to make the linkage between the level of evidence and grade of recommendation as transparent as possible.

RATING SCHEME FOR THE STRENGTH OF THE RECOMMENDATIONS

Grades of Recommendation

A. Based on clinical studies of good quality and consistency addressing the specific recommendations and including at least one randomized trial
B. Based on well-conducted clinical studies, but without randomized clinical studies
C. Made despite the absence of directly applicable clinical studies of good quality

COST ANALYSIS

A formal cost analysis was not performed and published cost analyses were not reviewed.

METHOD OF GUIDELINE VALIDATION

Internal Peer Review

DESCRIPTION OF METHOD OF GUIDELINE VALIDATION

There is no formal external review prior to publication.

The Appraisal of Guidelines for Research and Evaluation (AGREE) instrument was used to analyse and assess a range of specific attributes contributing to the validity of a specific clinical guideline.

The AGREE instrument, to be used by two to four appraisers, was developed by the AGREE collaboration (www.agreecollaboration.org) using referenced sources for the evaluation of specific guidelines. (See the "Availability of Companion Documents" field for further methodology information).

RECOMMENDATIONS

MAJOR RECOMMENDATIONS

Diagnosis

The challenge in the management of dilated upper urinary tracts is to decide which child can be observed, which one can be managed medically, and which one requires surgical intervention. There is no single diagnostic test that is definitive for distinguishing obstructive from non-obstructive cases (see Figure 2 in the original guideline document).

Antenatal Ultrasound

The kidneys are usually visualized routinely between the 16th and 18th weeks of pregnancy, when almost all amniotic fluid consists of urine. The 28th week is the most sensitive time for fetal urinary tract evaluation. If dilatation is detected, ultrasound should focus on the laterality, severity of dilatation and echogenicity of the kidneys, hydronephrosis or hydro-ureteronephrosis, bladder volume and bladder emptying, sex of the child and amniotic fluid volume.

Postnatal Ultrasound

Because transitory neonatal dehydration lasts about 48 hours, imaging should be performed after this period of postnatal oliguria. Immediate postnatal sonography is recommended in severe cases (bilateral dilatation, solitary kidney, oligohydramnios). During ultrasound examination, the anteroposterior diameter of the renal pelvis, calyceal dilatation, kidney size, parenchymal thickness, cortical echogenicity, ureters, bladder wall and residual urine are assessed.

Voiding Cystourethrogram (VCUG)

In newborns with identified upper urinary tract dilatation, the presence of primary or important associated factors that must be detected include vesicoureteral reflux in up to 25% of affected children, urethral valves, ureteroceles, diverticula and neurogenic bladder. Conventional VCUG is the method of choice for primary diagnostic procedures.

Diuretic Renography

Diuretic renography is the most commonly used diagnostic tool to detect the severity and functional significance of urine transport problems. Technetium-99m mercaptoacetyltriglycine (99mTc-MAG3) is the radionuclide of choice. It is important to perform the study under standardized circumstances (hydration, transurethral catheter) between the fourth and sixth weeks of life.

Oral fluid intake is encouraged prior to the examination. Fifteen minutes before the injection of the radionuclide, normal saline intravenous infusion at a rate of 15 mL/kg over 30 minutes is mandatory, with a subsequent maintenance rate of 4 mL/kg/h during the whole time of the investigation. The recommended dose of furosemide is 1 mg/kg for infants during the first year of life, while 0.5 mg/kg should be given to children aged 1 to 16 years up to a maximum dose of 40 mg.

Treatment

Prenatal Management

Counselling the parents is one of the most important aspects of care. The prognosis for an hydronephrotic kidney, even if severely affected, is hopeful. An hydronephrotic kidney may still be capable of delivering meaningful renal function in contrast to a severely hypoplastic and dysplastic kidney which has a hopeless outlook.

It is important to explain to the parents the timing and accuracy of establishing the definitive diagnosis for their child. In some cases, there is an obvious indication of severity, including massive bilateral dilatation, bilateral evidence of hypoplastic dysplasia, progressive bilateral dilatation with oligohydramnios and pulmonary hypoplasia.

Intrauterine intervention is rarely indicated and should only be performed in well-experienced centres.

Ureteropelvic Junction (UPJ) Obstruction

It is most important to make the decision on the basis of serial investigations applying the same technique and performed by the same institution under standardized circumstances. Symptomatic obstruction (recurrent flank pain, urinary tract infection) requires surgical correction using a pyeloplasty, according to the standardized open technique of Hynes and Anderson. In asymptomatic cases, conservative follow-up can be the treatment of choice.

Indications for surgical intervention comprise an impaired split renal function (less than 40%), a decrease of split renal function of more than 10% in subsequent studies, increased anteroposterior diameter on the ultrasound, and grade III and IV dilatation as defined by the Society for Fetal Urology.

Megaureter

Concerning the treatment options of secondary megaureters, (see reflux & valves, in the National Guideline Clearinghouse (NGC) summary of the EAU guideline, vesicoureteric reflux [VUR]). If a functional study reveals and confirms adequate

ureteral drainage, conservative management is the best option. Initially, low-dose prophylactic antibiotics within the first year of life are recommended for the prevention of urinary tract infections, although there are no existing prospective randomized trials evaluating the benefit of this regimen.

With spontaneous remission rates of up to 85% in primary megaureter cases, surgical management is no longer recommended except for megaureters with recurrent urinary tract infections, deterioration of split renal function and significant obstruction.

The initial approach to the ureter can be either intravesical, extravesical or combined. Straightening the ureter is necessary without devascularization. Ureteral tapering should enhance urinary flow into the bladder. The ureter must be tapered to achieve a diameter for an antireflux repair. Several tailoring techniques exist, such as ureteral imbrication or excisional tapering.

Conclusion

With the use of routine perinatal sonography, hydronephrosis caused by UPJ or ureterovesical junction (UVJ) obstruction is now recognized in increasing numbers. Meticulous and repeat postnatal evaluation is mandatory to try to identify those obstructive cases at risk of renal deterioration and requiring surgical reconstruction. Surgical methods are quite standardized and have a good clinical outcome.

CLINICAL ALGORITHM(S)

The original guideline document contains a diagnostic clinical algorithm for dilatation of the upper urinary tract.

EVIDENCE SUPPORTING THE RECOMMENDATIONS

TYPE OF EVIDENCE SUPPORTING THE RECOMMENDATIONS

The type of supporting evidence is not specifically stated or each recommendation.

BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS

POTENTIAL BENEFITS

- Appropriate diagnosis, treatment, and management of dilatation of the upper urinary tract in children
- Restoration of normal urinary flow
- Prevention of deterioration of the kidney

POTENTIAL HARMS

Not stated

QUALIFYING STATEMENTS

QUALIFYING STATEMENTS

The purpose of these texts is not to be proscriptive in the way a clinician should treat a patient but rather to provide access to the best contemporaneous consensus view on the most appropriate management currently available. European Association of Urology (EAU) guidelines are not meant to be legal documents but are produced with the ultimate aim to help urologists with their day-to-day practice.

IMPLEMENTATION OF THE GUIDELINE

DESCRIPTION OF IMPLEMENTATION STRATEGY

The European Association of Urology (EAU) Guidelines long version (containing all 19 guidelines) is reprinted annually in one book. Each text is dated. This means that if the latest edition of the book is read, one will know that this is the most updated version available. The same text is also made available on a CD (with hyperlinks to PubMed for most references) and posted on the EAU websites Uroweb and Urosource (www.uroweb.org/professional-resources/guidelines/ & http://www.urosource.com/diseases/).

Condensed pocket versions, containing mainly flow-charts and summaries, are also printed annually. All these publications are distributed free of charge to all (more than 10,000) members of the Association. Abridged versions of the guidelines are published in European Urology as original papers. Furthermore, many important websites list links to the relevant EAU guidelines sections on the association websites and all, or individual, guidelines have been translated to some 15 languages.

IMPLEMENTATION TOOLS

Clinical Algorithm

For information about availability, see the "Availability of Companion Documents" and "Patient Resources" fields below.

INSTITUTE OF MEDICINE (IOM) NATIONAL HEALTHCARE QUALITY REPORT CATEGORIES

IOM CARE NEED

Getting Better
Living with Illness

IOM DOMAIN

Effectiveness
Patient-centeredness

IDENTIFYING INFORMATION AND AVAILABILITY

BIBLIOGRAPHIC SOURCE(S)

Dilatation of the upper urinary tract (ureteropelvic junction and ureterovesical junction obstruction). In: Tekgul S, Riedmiller H, Gerharz E, Hoebeke P, Kocvara R, Nijman R, Radmayr C, Stein R. Guidelines on paediatric urology. Arnhem, The Netherlands: European Association of Urology, European Society for Paediatric Urology; 2008 Mar. p. 44-7. [15 references]

ADAPTATION

Not applicable: The guideline was not adapted from another source.

DATE RELEASED

2008 Mar

GUIDELINE DEVELOPER(S)

European Association of Urology - Medical Specialty Society
European Society for Paediatric Urology - Medical Specialty Society

SOURCE(S) OF FUNDING

European Association of Urology

GUIDELINE COMMITTEE

Not stated

COMPOSITION OF GROUP THAT AUTHORED THE GUIDELINE

Primary Authors: S. Tekgül; H. Riedmiller; E. Gerharz; P. Hoebeke; R. Kocvara; R. Nijman; Chr. Radmayr; R. Stein

FINANCIAL DISCLOSURES/CONFLICTS OF INTEREST

All members of the working group submit a conflict of interest form. The information is kept on file in the European Association of Urology (EAU) Central Office database. This guidelines document was developed with the financial support of the EAU. No external sources of funding and support have been involved. The EAU is a non-profit organisation and funding is limited to administrative assistance, travel, and meeting expenses. No honoraria or other reimbursements have been provided.

GUIDELINE STATUS

This is the current release of the guideline.

GUIDELINE AVAILABILITY

Electronic copies: Available in Portable Document Format (PDF) from the European Association of Urology Web site.

Print copies: Available from the European Association of Urology, PO Box 30016, NL-6803, AA ARNHEM, The Netherlands.

AVAILABILITY OF COMPANION DOCUMENTS

The following are available:

- EAU guidelines office template. Arnhem, The Netherlands: European Association of Urology (EAU); 2007. 4 p.
- The European Association of Urology (EAU) guidelines methodology: a critical evaluation. Arnhem, The Netherlands: European Association of Urology (EAU); 18 p.

Print copies: Available from the European Association of Urology, PO Box 30016, NL-6803, AA ARNHEM, The Netherlands.

PATIENT RESOURCES

None available

NGC STATUS

This NGC summary was completed by ECRI Institute on November 17, 2008. The information was verified by the guideline developer on December 19, 2008.

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